axillae with the usual discoloration but only to a moderate degree. By that time, however, the diagnosis was clear cut. The patient died November 22, 1945, about five months after the first verrucae were noted.

### NECROPSY

Necropsy showed essentially generalized metastatic carcinoma. There was a carcinomatous infiltration of the stomach of the leather bottle type. The pathologist expressed the opinion that this was the primary seat of the tumor. It was of particular interest to note that despite the very diffuse metastasis, the lungs, kidneys, liver and spleen showed little involvement, whereas the left adrenal and practically all the abdominal lymph nodes showed marked carcinomatous infiltration. The left adrenal was markedly enlarged, measuring 5 by 3 by 3 centimeters. The central portion was composed of tumor tissue which destroyed the greater portion of the cortex.

### COMMENT

This case was of unusual interest to us because of the rapid diffuse eruption of discrete verrucous papules over practically the entire body prior to any appreciable involvement in the classical areas such as the neck, groin, or axillae. Later, these areas showed the typical verrucous, discolored appearance, but only to a moderate degree. Also unusual was the clinical course of events in that the whole development was seemingly very explosive. Even when seen by us with a rather profuse eruption the patient was still obese despite the "leather bottle stomach" and extensive metastasis. She did not seem very ill, nor was there much complaint with reference to internal organs. Any deductions would be of a speculative character, but the course of events and the necropsy findings would seem to indicate that the primary seat of the carcinomatous change was in the stomach, and that there it involved chiefly the glandular elements. When metastases did occur, they rather early affected those factors responsible for the development of the syndrome called acanthosis nigricans. In the further spread of the metastasis again there was almost a selective involvement of lymph nodes. The cachexia of metastatic carcinoma never developed. Even at autopsy the body was reported as well nourished. This case illustrates again that a close cooperation between the dermatologist and general clinician is desirable.

727 West Seventh Street.

*Discussion by* ERNEST K. STRATTON, M.D., *San Francisco*

Inasmuch as I haven't seen a case of acanthosis nigricans for many years, I will make my comments brief and limit them to one phase of the hypothetical aspects of its etiology.

The etiology and pathogenesis of acanthosis nigricans are far from being understood. The fact is that some cases of the adult form (that is, the malignant type which is usually associated with abdominal carcinoma) are reported in patients in whom no evidence of malignancy is found. It is also a fact that the juvenile form (that is, the benign type in which it is rare to find an associated malignancy) is occasionally reported associated with malignancy. These observations would seem to indicate that some other factor besides the malignancy itself is responsible for the skin picture, especially when the pathological changes in the skin are the same in both types.

One sign, namely the pigmentation, was believed by Darier and others to be caused by a disturbance of the nerve supply to the adrenal glands. The splanchnic nerves, a division of the abdominal autonomic system which innervate the gland cells of the medulla, are believed to act as inhibitors of pigmentation. When this function is interfered with by pressure, either by neoplasms of the abdominal viscera or by abdominal metastasic growths, as seen in the adult cases, or by the pressure of such conditions as congenital malformations, peritoneal adhesions and benign growths, as seen in the juvenile cases, paralysis of these nerves results and a hyperpigmentation follows.

Even if the pigmentation could be explained with such a theory, how could one account for the hyperkeratosis, the acanthosis and other aspects of the syndrome? Avitaminosis associated with malignancy might be one explanation. (As in Hollander's case).

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## Ruptured Mucocele of the Appendix With Pseudomyxoma Peritonei, Simulating Pseudomucinous Cystadenoma of the Ovary

MELFORD B. JORGENSEN, M.D., *Los Gatos*

THIS is the report of a case of ruptured mucocele of the appendix with marked generalized pseudomyxoma peritonei, simulating that associated with pseudomucinous cystadenoma of the ovary. This case is remarkable in that the mucus, instead of being confined to the vicinity of the appendix and cecum in a mass, was found throughout the abdominal cavity in large amounts.

Foot describes the mucocele as arising in an appendix that has been acutely inflamed, with cicatrization causing proximal occlusion and the sealed off distal part turning into a simple pouch. Mucus continues to be secreted into this pouch, and the secretion becomes inspissated and gelatinoid. The pouch, becoming distended, ruptures and its contents spill into the peritoneal cavity with resulting production of numerous deposits of mucus secreting cells that resemble those of pseudomucinous adenomas of the ovary. Although appendiceal mucoceles are usually not very large, some have been reported as enormous.

In the presence of mucus deposits the peritoneum usually shows signs of reaction to irritation. A fine pellicle of fibrin forms over the mucoid material, and fine fibrous septa develop, which at times makes removal difficult.

### CASE REPORT

The patient, a 62-year-old white married female, was first seen because of complaint of epigastric distress and soreness in the lower abdomen on the right side of one week's duration. The patient felt nauseated but did not vomit. She was constipated, had poor appetite and said that she had been very nervous and in the past year had been given considerable sedatives. There was no history of operations or of serious illness or injury.

The skin and mucous membranes seemed somewhat drier than normal. The temperature was 99.2° F., the pulse rate was 90 and the blood pressure 180 mm. of mercury systolic, and 90 mm. diastolic. No cardiac abnormalities were noted. There were no palpable masses in the abdomen. Very slight rigidity was noted in the right lower quadrant of the abdomen and there was moderate tenderness in that area, but no rebound tenderness. Rectal examination revealed tenderness high on the right side. On pelvic examination the uterus was found to be retroverted but normal in size. Adnexa were normal. The cervix was slightly eroded.

Examination of the blood showed hemoglobin 81.5 per cent. Erythrocytes numbered 3.93 million and leukocytes 7,400 with normal distribution of polymorphonuclear cells,

lymphocytes, stabs and segmented cells. A Wassermann test was negative. Results of urinalysis were normal.

At operation the peritoneum and serosal surface of the small and large bowel and pelvic viscera were injected and covered with a fibrinous membrane. There was a large amount of gelatinous material present throughout, but mainly located in both lower quadrants, deep in the pelvis. A large mass of this substance was present in the right lower quadrant at the base of the cecum. The appendix 3 cm. from the cecum entered a globular mass measuring 6 x 3 cm. which was hard, and whitish in color. In the lower portion of it was a hole 1½ cm. in diameter, out of which exuded gelatinous material. This mass lay retrocecal and was bound down to the posterior aspect of the cecum by dense adhesions. Both ovaries were small, fibrotic, and intact. The Fallopian tubes and uterus appeared normal except for serosal injection as mentioned.

The appendix, with the mass, was removed, as was the gelatinous material from the rest of the abdomen.

*Pathological Report:* The gross specimen was a tubular mass 5.2 cm. long and 2.4 cm. wide. It was closed at one end, which was blunt, and was clamped at the other end. The outer surface was opaque and consisted of friable, yellow-white tissue. The tube was slightly curved and in the concavity was a 2.4 x 2.2 cm. ovoid pouch or mass, gelatinous in consistency and covered with thin gray membrane. Examination of a section showed the tubular lumen to be dilated, the wall measuring 0.3 cm. in thickness. The lumen was continuous with a cavity in the gelatinous mass, both being filled with a gray, thick, mucoid semiliquid, which was intimately adherent to the walls. Calcium or bone was present in the wall, making it firm and gritty in certain areas.

Microscopic examination showed the serosa was intact, but the subserosal connective tissue was edematous and contained numerous small and large vascular channels which were uniformly dilated and filled with red blood corpuscles. Collections of lymphocytes about some of these vessels were noted. The muscular coats of the appendix were both notably thickened but otherwise were not remarkable. The mucosa was markedly altered, there being no evidence of the lymphoid follicles. The glandular structure was somewhat effaced, and the lining mucosal cells were thrown into papillary folds and rested directly upon the described musculature. The cells composing the altered mucosa were tall and columnar, with basally placed, elongated, hyperchromatic nuclei and with overriding cytoplasm that was distended with a neutrophilic-staining material, probably mucoid.

When sections through the appendiceal wall and also through the mass that was described in its concavity were examined, the mass appeared to be continuous with the appendiceal mucosa and it consisted for the most part of irregular strings and strands and small collections of mucoid material which was neutrophilic-staining and which in some places was collected and separated from other portions by lacy, thin fibrous connective tissue strands. A few fibrocytes were noted and there were a few areas resembling cartilage. In other portions the wall contained granular plaques of deep blue-staining material which appeared to be calcium. In one portion the small tumor was ruptured and the mucoid material was pouring out through the orifice. Malignant changes were not observed. Inflammatory reaction was not a feature.

*Diagnosis:* Mucocele, appendix. Pseudomyxoma peritonei.

The patient made an uneventful recovery and was dismissed from the hospital on the fifth postoperative day. She has been observed about once a month subsequently and there have been no complaints of abdominal distress.

## COMMENT

These rare cysts of the appendix are of interest to the surgeon because they may be the starting point of a condition of pseudomyxoma peritonei similar to that which occasionally complicates pseudomucinous cystadenomas of the ovary. Boyd states that as a result of rupture of the cyst, the contents become implanted on the surface of the peritoneum where they cause proliferation and the formation of large masses of gelatinoid material. The condition is probably due to implantation of the epithelial cells on the peritoneal surface, where they continue to produce their mucinous secretions. Another explanation is that the material irritates the peritoneum and causes it to react increasingly to further production of similar masses. At any rate, fibrous tissue proliferation in the peritoneum takes place, so that eventually it may be impossible to remove the masses.

The prognosis according to Boyd is unfavorable, but removal of the appendix may check the disease. The disease is fatal in the majority of cases, although a number of years may elapse between onset and death. Recurrence of the peritoneal reaction may be expected. Many of the patients die of sepsis and embolism.

Corner Massal and Saratoga Avenues.

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